

CONTINUING EDUCATION PROGRAM: FOCUS...

The lower cranial nerves: IX, X, XI, XII



J.-L. Sarrazin^{a,b,*}, F. Toulgoat^c, F. Benoudiba^b

^a Service d'imagerie médicale, American Hospital of Paris, 63, boulevard Victor-Hugo, 92200 Neuilly-sur-Seine, France

^b Service de neuroradiologie, CHU de Bicêtre, 78, rue du Général-Leclerc, 94270 Le Kremlin-Bicêtre, France

^c Neuroradiologie diagnostique et interventionnelle, CHU de Nantes, hôpital Laennec, boulevard Jacques-Monod–Saint-Herblain, 44093 Nantes cedex 1, France

KEYWORDS

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Meningioma

Abstract The lower cranial nerves innervate the pharynx and larynx by the glossopharyngeal (CN IX) and vagus (CN X) (mixed) nerves, and provide motor innervation of the muscles of the neck by the accessory nerve (CN XI) and the tongue by the hypoglossal nerve (CN XII). The symptomatology provoked by an anomaly is often discrete and rarely in the forefront. As with all cranial nerves, the context and clinical examinations, in case of suspicion of impairment of the lower cranial nerves, are determinant in guiding the imaging. In fact, the impairment may be located in the brain stem, in the peribulbar cisterns, in the foramina or even in the deep spaces of the face. The clinical localization of the probable seat of the lesion helps in choosing the adapted protocol in MRI and eventually completes it with a CT-scan. In the bulb, the intra-axial pathology is dominated by brain ischemia (in particular, with Wallenberg syndrome) and multiple sclerosis. Cisternal pathology is tumoral with two tumors, schwannoma and meningioma. The occurrence is much lower than in the cochleovestibular nerves as well as the leptomeningeal nerves (infectious, inflammatory or tumoral). Finally, foramen pathology is tumoral with, outside of the usual schwannomas and meningiomas, paragangliomas. For radiologists, fairly hesitant to explore these lower cranial pairs, it is necessary to be familiar with (or relearn) the anatomy, master the exploratory technique and be aware of the diagnostic possibilities.

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The lower cranial nerves run from the 3rd and 4th branchial (CN IX and CN X, respectively) motor, sensory and secretory arches and the somatic motor nerves for the latter two (CN XI and CN XII).

The symptomatology related to their impairment is often little marked and rarely at the forefront of patient complaints.

* Corresponding author. Service d'imagerie médicale, American Hospital of Paris, 63, boulevard Victor-Hugo, 92200 Neuilly-sur-Seine, France.

E-mail address: jlsarrazin@wanadoo.fr (J.-L. Sarrazin).

For this reason, radiologists are not often confronted with the exploration of these nerves and the relative difficulty of the anatomy makes them circumspect when faced with a request to examine these nerves.

The purpose of this article is therefore to review, in a simple manner, the anatomy, clinics, mode of exploration and pathology of these nerves.

Presentation of the nerves

The IXth pair of cranial nerves: the glossopharyngeal nerve

This is a mixed sensory, motor and secretory nerve (Table 1).

It is often synergic with the vagus nerve (CN X).

It ensures the sensory innervation of the pharynx. It is motor only for the stylopharyngeal muscle. It also has a secretory function by regulating the salivary secretion of the parotid glands.

The Xth pair of cranial nerves: the vagus nerve

This is a mixed sensory and motor nerve.

Along with CN IX, it provides part of the sensory and motor innervation of the pharynx.

It is the motor nerve of the larynx.

It is also the parasympathetic nerve of the thoracic and abdominal viscera.

The XIth pair of cranial nerves: the accessory nerve

This is mainly a motor nerve.

This nerve is unusual since it mainly consists of a spinal contingent that innervates the neck muscles (trapezoid and sternocleidomastoid or SCM) and an "accessory" contingent of CN X that innervates the larynx. Certain anatomists contest this cranial contingent of CN XI.

The XIIth pair of cranial nerves: the hypoglossal nerve

This is only a motor nerve.

It innervates all of the muscles in the tongue.

Anatomic approach [1–4]

Besides the first two pairs of cranial nerves, more prolongations of the brain than real nerves, all of the pairs of cranial nerves comply with the same plan:

- a nucleus (or nuclei) in the brain stem (except for CN XI);
- a cisternal portion;
- a foramen (or foramina) for exit from the skull;
- collateral branches and effector nerve endings.

The nuclei

For the IXth, Xth and XIIth pairs of cranial nerves (and the cranial contingent from CN XI)

The nuclei are found at the posterior part of the bulb in the region of the floor of the IVth ventricle.

The nucleus ambiguus (motor)

It is part of the branchial motor column of the brain stem. The most anterior and lateral, it gives rise to the motor fibers intended for the muscles of the pharynx and larynx. These fibers thereby participate in the formation of the glossopharyngeal nerve (CN IX), the vagus nerve (CN X) and very partially, the accessory nerve (CN XI).

The solitary nucleus (sensory)

It is part of the column of the efferent nuclei from the brain stem. It is more median than the solitary nucleus. It receives the sensory afferents from the pharynx and larynx, thereby helping form the glossopharyngeal nerve and vagus nerve.

The inferior salivary nucleus (secretory)

It controls the secretion of the salivary glands, in particular the parotid glands. Its fibers are conveyed by the glossopharyngeal nerve.

Table 1 The lower cranial nerves: anatomical summary.

Nerves	Nuclei	Emergence	Foramen	Effectors	Function
IX Glossopharyngeal	Ambiguus (M) Solitary (S) Inferior salivary (Secret.)	Retro-olivary groove	Jugular foramen	Pharynx S M (stylopharyngeal) Parotid	Synergic with X S and M for the pharynx
X Vagus	Ambiguus (M) Solitary (S) Dorsal (paraΣ)	Retro-olivary groove	Jugular foramen	Pharynx (M and S) Larynx (M) Viscera thorax and abdomen (paraΣ)	Pharynx (S and M) Larynx (M) Viscera (paraΣ)
XI Accessory	Ventromedial medullar C2 to C6	Lateral medullary cord	Jugular foramen	SCM Trapezium	Motor
XII Hypoglossal	Hypoglossal	Pre-olivary groove	Hypoglossal canal	Muscles of the tongue	Motor

M: motor; S: sensory; Secret.: secretory; ParaΣ: parasympathetic; SCM: sternocleidomastoid.

The hypoglossal nucleus (motor)

It is part of the column of somatic motor nuclei.

It is paramedian and gives the floor of the IVth ventricle an arch (hypoglossal eminence).

It provides all of the muscles of the tongue with motor efferences by forming the hypoglossal nerve.

For the spinal contingent of CN XI

The ventral nuclei of the anterior horn of the spinal cord (*motor*).

Their afferences form the spinal contingent of the accessory nerve to innervate the trapezius and sternocleidomastoid.

The cisternal portion**IXth, Xth pairs of cranial nerves**

They exit the bulb at the posterolateral groove of the spinal cord (retro-olivary groove). They then run relatively horizontally oblique forward and outside to reach the jugular foramen.

XIth pair of cranial nerves

The spinal contingent from CN XI advances in the anterior spinal subarachnoid spaces to enter the skull by the magnum foramen and associates with cranial nerves IX and X along their cisternal pathway.

XIIth pair of cranial nerves

The fibers exit the bulb from the ventrolateral groove of the bulb (pre-olivary groove) in the form of several rootlets, a little below the level of the previous pairs of cranial nerves.

Here too, their pathway is horizontally oblique forwards and outside.

Like most of the cranial nerves, the origin of the myelin of the cisternal portion of these nerves is double: near their emergence from the bulb, the myelin is oligodendrocytary, of central origin. Then, more laterally, the myelin becomes peripheral produced by Schwann cells. The zone of transition between the (more medial) portion of the nerve, where the myelin is central, and the more lateral portion, where the myelin is peripheral, is called the Root Entry Zone (REZ) and is a zone of nerve fragility.

The seat of this REZ is relatively constant for certain nerves (CN V, CN VII and CN VIII) and is more variable for others, as is true of the lower cranial nerves [3].

The foramina [5]

There are two foramina for these four nerves:

- the jugular foramen through which exit the glossopharyngeal nerve, the vagus nerve and the accessory nerve (CN IX, CN X, CN XI);
- the hypoglossal canal for the exit of the hypoglossal nerve (CN XII) from the skull.

The jugular foramen

It is a large foramen created at the lower side of the skull between the temporal bone and the occipital bone.

The main axis has an oblique orientation forward and inwards.

It is piriform with a posterior-lateral rounded part and a more anterior-medial slender part.

The posterior rounded part contains the superior bulb.

The slender part is divided into two by the petro-occipital ligament:

- the glossopharyngeal nerve passes in front and outside;
- the vagus and accessory nerves pass to the rear and inside as does the posterior meningeal artery.

One of the particularities of the jugular foramen is the presence of glomic bodies both opposite the superior bulb and especially cranial nerves X and XI. These glomic bodies give rise to the paragangliomas.

The hypoglossal canal

It is a small oblique canal in front of and outside that only allows the hypoglossal nerve surrounded by venous plexus to pass.

The extracranial branches

The pathway of the branches of CN XI is below and to the rear to be distributed to the trapezium and sternocleidomastoid.

The branches of CN IX, CN X and CN XII run in the retro-styloid space to reach the effector organs (the pharynx for CN IX, the pharynx, larynx, thorax and abdomen for CN X and the tongue for CN XII).

CN IX gives:

- the tympanic nerve that provides the sensitivity of the eardrum and the auditory tube;
- the stylopharyngeal branch (motor);
- the pharyngeal branch (sensory) that associates to the fibers of CN X;
- the carotid sinus branch;
- the lingual branch for the taste of the posterior 1/3 of the tongue.

CN X gives:

- a dural branch for the dura mater;
- an auricular branch for the skin sensitivity of the posterior part of the auricle;
- the pharyngeal branches that associate with the fibres of CN IX;
- the superior laryngeal nerve is motor for the constrictors of the pharynx and sensory for the pharynx;
- the inferior laryngeal nerve is motor for the intrinsic musculature of the larynx;
- cardiac, bronchial and gastric branches.

CN XI and CN XII are efferent somatic nerves that do not give rise to collateral branches.

The close contact between CN XII and the carotid artery in the retro-styloid space should be noted.

Clinics

The glossopharyngeal nerve

Its impairment is rarely isolated. Most often, it is concomitant with that of the vagus nerve. A lesion of the glossopharyngeal nerve induces:

- aguesia in the posterior third of the tongue;
- abolition of the position of the vomiting and velopalatin reflexes;
- anesthesia of the upper part of the pharynx, the tonsils and the base of the tongue;
- minor difficulty swallowing.

However, there is specific syndrome for the glossopharyngeal nerve: glossopharyngeal neuralgia [6]:

- it consists of painful episodes similar to facial neuralgia, with paroxysmal pain, a sudden onset and relatively short episodes;
- most often, it begins at the base of the tongue and/or tonsils, or palate and radiates back towards the ear;
- there is a triggering factor (chewing, swallowing, coughing, speaking);
- in most cases, it is due to an arterial (posterior inferior cerebellar artery) – nerve (glossopharyngeal nerve) conflict.

The vagus nerve

Impairment of the vagus nerve is often associated with that of the glossopharyngeal nerve.

A complete unilateral lesion of the vagus nerve provokes:

- paralysis of the pharyngeal muscles with lowering of the palate on the impaired side and attraction of this palate and the uvula on the healthy side during phonation;
- paralysis of the larynx with paralysis of the homolateral vocal cord and a nasal voice;
- minor dysphagia may also exist, as well as tachycardia and arrhythmia.

The accessory nerve

Impairment is rare and will induce paralysis of the sternocleidomastoid (SCM) and trapezius. In this case, the paralysis of the SCM is limp and complete while that of the trapezius only involves the upper part of the muscle. Clinically, the patient presents:

- a lowering of the shoulder related to the impairment of the trapezius;
- a reduction in the relief of the neck and is unable to turn his head on the healthy side due to paralysis of the SCM muscle.

The hypoglossal nerve

Due to the paramedian location of the right and left nuclei of CN XII, nuclear impairment is most often bilateral. The impairment is unilateral downstream.

In this case, there will be lingual hemiatrophy, with a seemingly crenate tongue.

When the tongue is protracted, it is deviated on the paralyzed side.

Syndrome study

Pseudobulbar paralysis, by bilateral impairment of the corticobulbar tract of vascular origin, will provoke spastic paresis of the muscles innervated by the lower cranial nerves. The patient will also be subject to pathological laughing and crying.

Progressive bulbar paralysis will be manifested by dysarthria, swallowing difficulties, atrophy and fasciculation of the tongue, followed by the appearance of nystagmus, ptosis and facial paresis. It occurs following amyotrophic lateral sclerosis syringomyelia.

The jugular foramen syndrome associates phonation disorders (hoarseness), swallowing disorders, regurgitation of liquids through the nose, sometimes excess salivation and coughing. The examination reveals paralysis of the superior pharyngeal constrictor, constant Vernet's rideau phenomenon (lowering of the soft palate on the paralyzed side), paralysis of the vocal cords, sternocleidomastoid, trapezius, aguesia of the posterior part of the tongue and hemianesthesia of the palate, pharynx and larynx.

Wallenberg syndrome, by occlusion of the posterior inferior cerebellar artery, often due to a dissection of the vertebral artery, associates difficulties swallowing and dysphonia (ambiguous nucleus), sensory disorders on the face, vertigo, and a cerebellar syndrome on the impaired side and thermoalgie hemianesthesia respecting the face on the impaired side.

Collet-Sicard syndrome, often related to a dissection or fracture of the base of the skull, associates impairment of the lower cranial nerves without sympathetic impairment.

Villaret's syndrome associates the same impairment of the lower cranial nerves and sympathetic impairment. The retro-styloid space is the seat of the lesion.

Exploration technique

MRI is the key examination to explore the cranial nerves. The CT-scan remains a highly useful complementary examination in the foraminal exploration to assess the type of bone impairment in case of a tumor.

The exploration technique should be guided by the clinics. In fact, a suspicion of central "nuclear" impairment is not explored in the same way as a suspicion of impairment of the cisternal part of the nerve or impairment of the effector branches in the deep spaces of the face.

Central impairment

The three basic sequences are:

- a FLAIR sequence (axial or better still, volume);
- a diffusion sequence, if necessary optimized for the infratentorial space (fine slices, tensor, spin echo acquisition);
- a susceptibility sequence ("classic" T2*-weighted or SWI (susceptibility weighted imaging)).

With the slightest doubt, a spin echo T2-weighted sequence will be acquired in fine slices, as this is especially effective in detecting signal anomalies in the case of multiple sclerosis, for example. Depending on the results of these sequences, the exploration will be completed:

- in case of suspicion of vascular impairment, by a 3D TOF MR angiography, more or less a T1-weighted sequence with fat saturation on the neck (or T1-weighted volume acquisition with fat saturation) in the search for a dissection;
- in case of suspicion of tumoral or inflammatory impairment, T1-weighted sequences without and then after injection of contrast product in fine slices.

Cisternal and foraminal impairment [7,8]

The basic sequence is a high-resolution T2-weighted sequence (or with T2 effect) such as Fiesta, Ciss, Drive according to the manufacturers.

As a rule, it is completed with T1-weighted sequences without and then after the injection of contrast product in fine slices.

The exploration also includes a full brain exploration, as a rule a FLAIR sequence and a diffusion sequence.

Impairment of the nerves and their extracranial branches

This involves the exploration of the deep spaces of the face that requires centered sequences with a good spatial resolution (fine slices, small field of vision, relatively extensive matrix), T2-weighted with fat saturation in axial and then orthogonal plane, T1-weighted with fat saturation and T1-weighted after injection and with fat saturation. A T1-weighted volume acquisition (LAVA or Vibe) after injection and with fat saturation may also be acquired as a complement.

In case of the presence of artifacts of dental origin, the use of multicontrast sequences (Dixon, Ideal) is indicated.

Pathology and imaging

Intra-axial pathology

Tumor pathology

It is rare in the adult (Table 2).

Gliomas of the trunk are most often low grade and symptomatology of impairment of the lower cranial nerves is exceptionally detected.

Table 2 The lower cranial nerves: clinical summary and technique.

Seat	Nerve(s)	Etiology	Technique
Supratentorial level Pseudobulbar palsy	Association IX, X, XI, XII ± VII	Vascular Bilateral VCA Bilateral tumors SLA	Flair T2*-weighted diffusion ± TOF/injection
Brain stem	Association IX, X, XI, XI Isolated impairment or by 2	SLA, polio Vascular SEP Infection	Diffusion T2*-weighted Fine slices T2-weighted ± injection
Cistern	IX IX, X, XI, XI	Neuralgia Meta, infection Tumor Schwannoma Meningioma	T2-weighted HR ± TOF ± T1-weighted without and with injection
Foramen	IX, X, XI XII XI	Meta Tumor Paraganglioma Schwannoma Meningioma Schwannoma Meningioma Foramen Magnum	Trauma T2-weighted T1-weighted without and with injection ± fat sat
Retro-styloid space neck	IX, X, XI, XII XI XII, CBH	Tumor Schwannoma Kc ORL Trauma, surgery Dissection	T2-weighted fat sat T1-weighted without T1-weighted with fat sat T1-weighted fat sat, MR angio

In case of impairment of the lower cranial nerves, tumoral pathology is highly exceptional. A case of exophytic pilocytic astrocytoma with exclusive impairment of the cranial nerves is one of these exceptional cases [9].

Vascular pathology

Nuclear impairment may be the result of ischemia. Wallenberg syndrome, by occlusion of the artery of the occlusion of the posterior inferior cerebellar artery, is the result of ischemia affecting the nuclei:

- of CN V with sensory disorders of the hemiface;
- vestibular with vertigo;
- ambiguous and solitary with pharyngeal paralysis.

There is cerebellar impairment with impairment of the homolateral inferior cerebellar peduncle and contralateral thermoalgie hemianesthesia with ischemia by impairment of the lemniscus pathway. This occlusion is often due to a dissection of the vertebral artery.

In the imaging, in view of an acute picture of alternate symptomatology, an optimized diffusion sequence for the brain stem should be acquired. It shows a more or less extensive intense lateral medullary lesion with restriction of the apparent diffusion coefficient. A T2-weighted sequence in

fine slices, FLAIR (volume) and a MR angiography complete the assessment.

A search should be carried out for dissection of the vertebral artery by a T1-weighted sequence with fat saturation (at best with a volume sequence with T1-weighted echo spin with fat saturation) and by an injected MR angiography (Fig. 1).

There may be hemorrhages of the brain stem in a hyper-intensive micro-angiopathy. However, here too the elective impairment of the lower cranial nerves at the forefront of the clinical picture is very rare.

Finally, it is necessary to mention the cavernous hemangiomas that are as a rule asymptomatic and may provoke bulbar nuclear impairment in case of hemorrhagic complications.

Inflammatory and infectious pathology

A demyelinating lesion may affect the bulb and the nuclei of the lower cranial nerves in cases of multiple sclerosis (Fig. 2).

In the imaging, the search is best carried out by fine slices with T2-weighting. The sensitivity of the volume FLAIR sequence is much higher than that of the 2D FLAIR in the detection of bulbar plaques, although it remains slightly

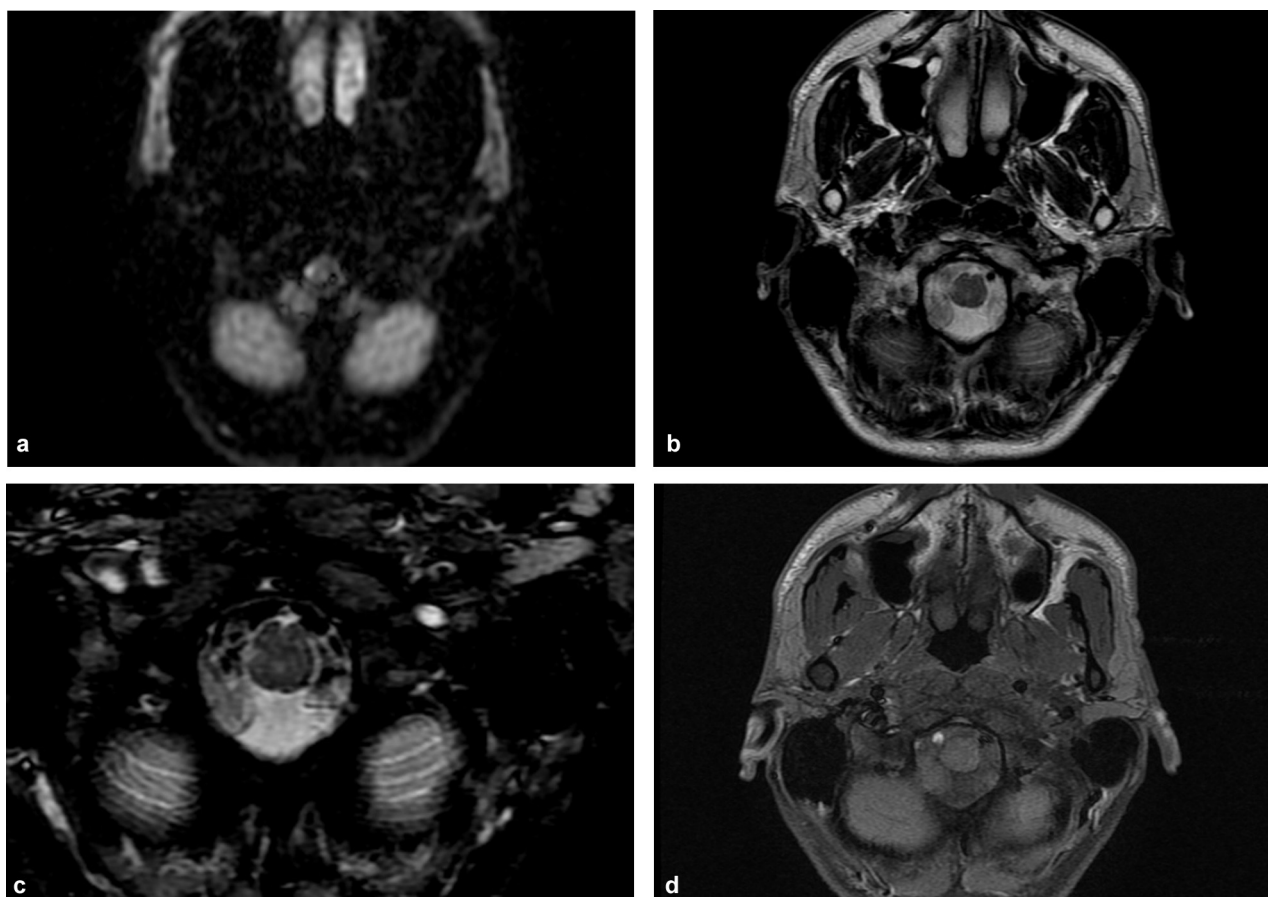


Figure 1. 39-year-old man. Sudden onset, the evening before, of vertigo with minor difficulty swallowing: a: diffusion imaging; b: T2-weighted axial slice; c: STIR axial slice; d: T1-weighted axial slice with fat saturation. Small, right laterobulbar lesion, intense in diffusion, slightly visible in the T2-weighted and STIR sequences with enlargement of the right vertebral artery by a parietal hematoma that blocks the arterial lumen. Wallenberg syndrome on vertebral dissection.

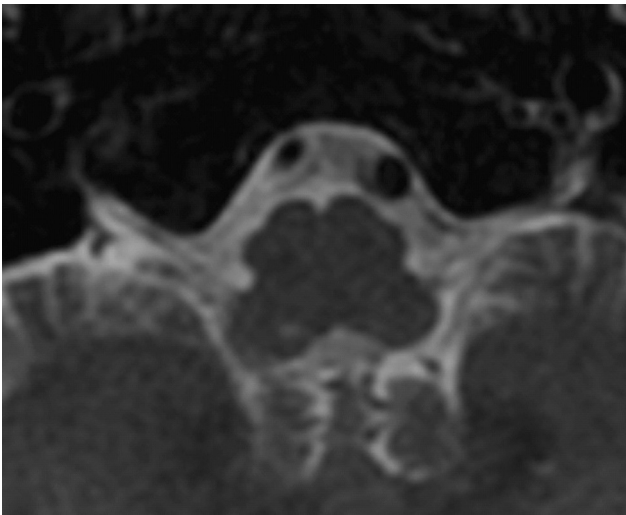


Figure 2. 63-year-old man. Antecedent of right endobuccal herpes zoster six months before. Persistence of dysguesia of the posterior part of the right hemitongue. T2-weighted axial slices on the bulb trunk. Small zoster lesion located on the solitary nucleus (sensory of CN IX).

inferior the T2-weighted sequences acquired in fine slices (Fig. 3).

Among the infectious impairments of the bulb, it's necessary to mention listeriosis and tuberculosis that may affect the nuclei of the lower cranial nerves:

- listeriosis, due to a bacterium, *Listeria monocytogenes*, occurs in weak patients with a symptomatology including coma, fever and impairment of the pairs of cranial nerves. The latter is probably due to a lesion of the nuclei resulting from inaugural meningitis by axonal dissemination. In the imaging, the signs of

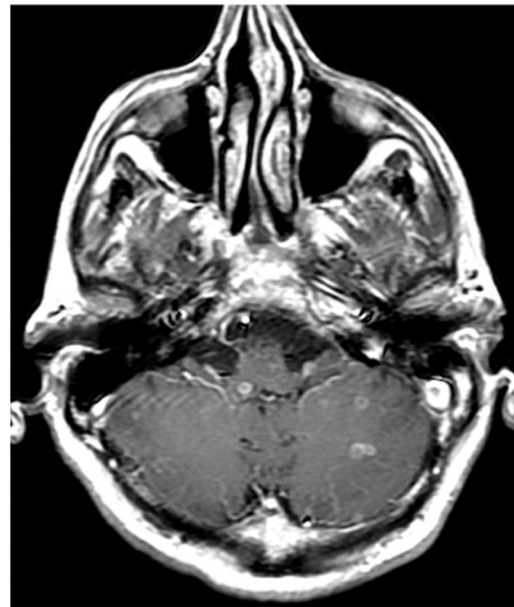


Figure 4. T1-weighted injected axial slice. Presence of multiple tuberculoma including one opposite the nuclei of the lower cranial nerves.

meningitis are often discrete. There are small, enhanced nodular formations after the injection of the contrast product located in the brain stem opposite nuclear clusters;

- tuberculosis: besides basilar meningitis, tuberculomes may disseminate in the brain stem. In the imaging, they appear in the form of small nodules, with a hypointense centre in T2, also rather hypointense in diffusion with peripheral enhancement (Fig. 4).

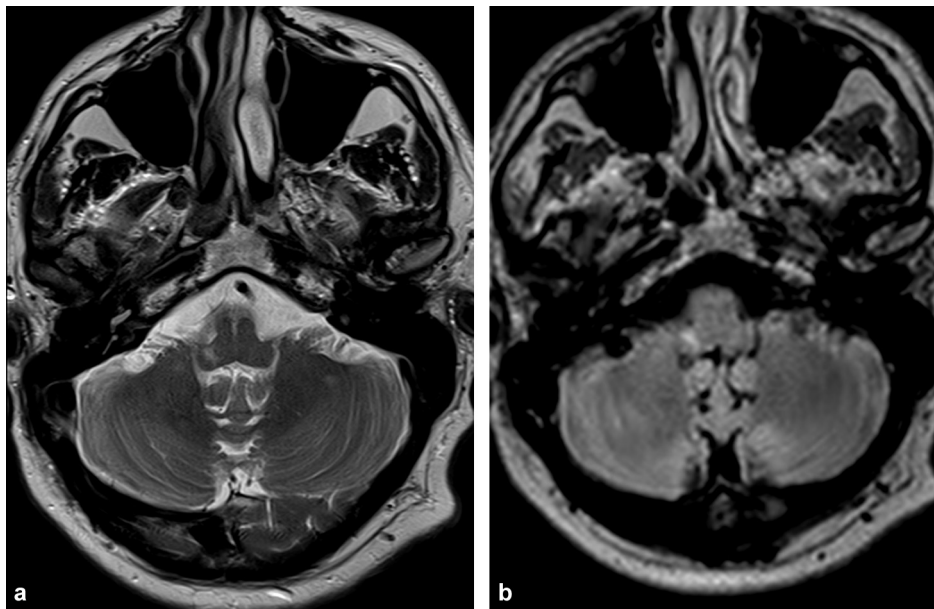


Figure 3. 39-year-old man. Known SEP. Appearance of sensory disorders of the base of the tongue and right tonsils: a: T2-weighted slices; b: axial-reconstructed FLAIR Volume acquisition. Plaque of the right part of the bulb affecting the nuclei of CN IX and CN X.

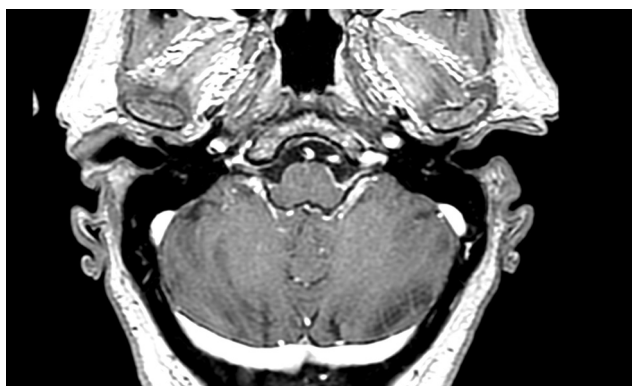


Figure 5. 64-year-old man. Bronchial cancer. Impaired consciousness. Impairment of the pairs of cranial nerves (III, V, VIII, IX, X). T1-weighted injected axial slice. Enhancement on the left of the leptomeninges of the bulb with extension to CN IX and X° left pairs of cranial nerves.

Cisternal pathology

Tumor pathology

Cisternal tumor pathology is, besides leptomeningeal metastases, benign mainly with schwannoma and meningioma. These tumors will be dealt with along with foraminal tumor pathology.

Due to the small size of the lower cranial nerves, often in the form of rootlets, their impairment during a malignant leptomeningeal dissemination is often difficult to detect and barely symptomatic. This impairment may be detected in lymphomatous patients (Fig. 5).

Inflammatory and infectious disease

Granulomatous disorders, in particular sarcoidosis, may affect the cisterns of the base.

Among the infections likely to affect the cranial nerves by leptomeningeal impairment, it's necessary to mention tuberculosis, neuroborreliosis and, in the immunosuppressed patient, cytomegalovirus and varicella zoster virus [10].

The artery-nerve conflict: glossopharyngeal neuralgia

The symptomatology has been noted above in the clinical section. It is necessary to note that this entity is often unknown, even by clinicians. In the imaging, to detect this conflict, the most pertinent sequence is the high-resolution T2-weighted sequence. It helps reveal the contact between the glossopharyngeal nerve and an artery, most often the posterior inferior cerebellar artery. The minIP post treatment sometimes helps improve the visualization of the conflict.

It should be noted that the exact seat of the REZ of the glossopharyngeal nerve is not determined and is probably variable. Moreover, the criteria of the conflict in the imaging of glossopharyngeal neuralgia are not as well defined as those for facial neuralgia. However, the right angle crossing and, above all, the shift of the nerve by the artery are of great diagnostic value.

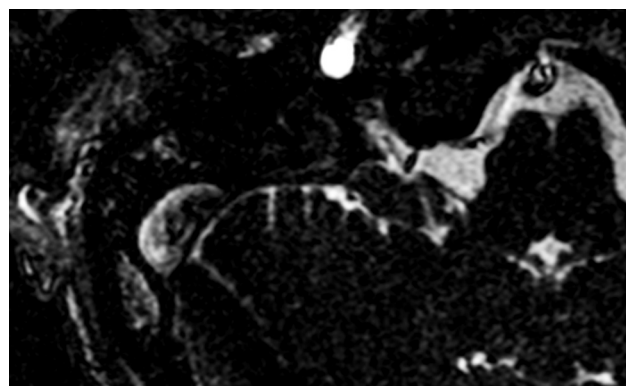


Figure 6. 33-year-old woman. Pain during meals of the right mandibular angle, extending to the ear. A parotid assessment looking for salivary colic was carried out and was normal. High-resolution T2-weighted slice. The right glossopharyngeal nerve is pushed back in its cisternal pathway by the posterior inferior cerebellar artery. Artery-nerve conflict. The patient improved with carbamazepine.

Other sequences (volume T1-weighted gradient echo, TOF MR angiography) are complementary sequences and eliminate other diagnoses (Fig. 6).

Foraminal pathology

It is rarely traumatic but rather tumoral. Only the tumoral pathology is dealt with in this section [11].

Three types of tumors have to be taken into account: the paragangliomas, the schwannomas and the meningiomas.

The schwannomas and meningiomas may also arise in the cisterns. The schwannomas may also be found in the retro-styloid space. Therefore, all three tumors may arise in the foramina and extend upwards in the cisterns and downwards in the retro-styloid space.

The paragangliomas [5,12,13]

The paragangliomas are tumors arising from glomic cells cervically located in the carotid bifurcation and along the vagus nerve and at the base of the skull in the jugular foramen and in the eardrum between the branches of Jacobson's nerve on the promontory (Fig. 7).

Histologically, it consists of an irregular, red mass, consisting of highly abundant epitheloid cells with a granular cytoplasm separated by a great many vessels. It comprises cells derived from the neural crest.

The occurrence of these tumors is low, 0.6% of the cervicocephalic tumors, one third of which are found in the tympanojugular.

Clinically, the main symptoms leading to the discovery of a paraganglioma are auditory, in particular, with pulsatile tinnitus and hypoacusia.

The jugular foramen syndrome that associates dysphagia, dysphonia (bitonal voice), dysarthria, hemiageusia of the posterior third of the tongue as well as paralysis of the sternocleidomastoid and the trapezius is more rare.

In the imaging, it consists of a mass that may be large, with irregular and poly-lobed contours, centered on

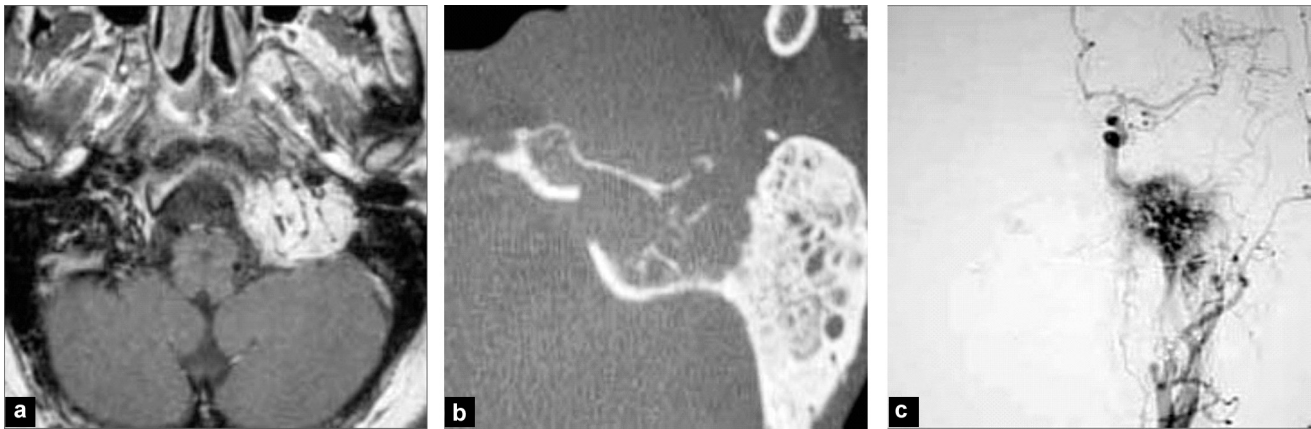


Figure 7. 53-year-old woman. Rather pulsatile left tinnitus. Minor difficulties swallowing: a: T1-weighted injected axial slice; b: CT-scan slice; c: arteriography. Mass centered on the left jugular foramen, highly enhanced after injection of contrast product with irregular contours, accompanied by “aggressive” lysis in CT-scan and a tumoral arterial blush in arteriography. Paraganglioma.

the jugular foramen. It may extend upwards towards the eardrum and, in particular the hypo-eardrum and more rarely downwards in the retro-styloid space.

Its signal is heterogeneous, rather hypointense in T1 and hyperintense in T2. The main characteristic is enhancement of its signal after injection, intense enhancement with the presence of tubular signal void structures within.

The presence of these intratumoral vessels associated with the intensity of the enhancement attests to the very vascular nature of this mass and supports the diagnosis of paraganglioma.

The CT-scan helps in the diagnosis by revealing an aggressive lesion with irregular lysis at the edges of the foramen, without condensing reaction.

An arteriography may be carried out before surgery and is also fairly characteristic of paraganglioma by revealing an arterial tumor blush.

The diagnosis with the two other tumors of the jugular foramen (schwannoma and meningioma) is, as a rule, easy when based on the morphological criteria and enhancement in the MRI and CT-scan.

The discovery of a paraganglioma requires a full cervical exploration to look for another location within a familial form. However, in these familial forms, the locations of the paragangliomas are more cervical than at the base of the skull.

As regards therapy, surgery is indicated. A pre-surgical arteriography with embolization may be discussed. However, it should be noted that full excision is often difficult due to the size of the tumor and the fact that there is very often a microscopic invasion of the cranial nerves, leading to recurrences even when the surgery seems to be macroscopically complete.

For this reason, complementary, targeted radiation therapy, whatever the form, is often carried out (gamma knife) [14].

The schwannomas [15]

Histologically, the schwannomas of the lower cranial nerves do not differ from vestibular cellular schwannomas (Antoni

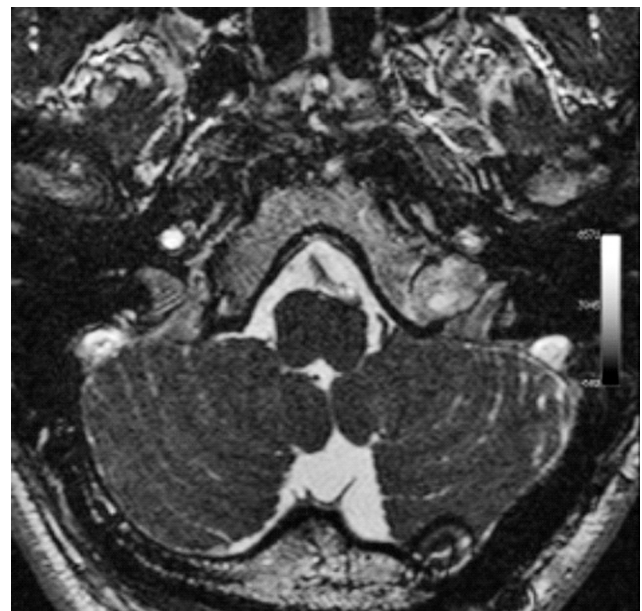


Figure 8. 38-year-old man. Mild difficulties swallowing. High-resolution T2-weighted axial slice after injection. Round mass centered on the left jugular foramen with small necrotic portions and accompanied by an even enlargement of the jugular foramen. Schwannoma of CN IX.

A) and myxoid schwannomas (Antoni B). Schwannomas of CN IX are more frequent (Fig. 8).

Clinically, schwannomas of the jugular foramen appear as paragangliomas, especially by auditory symptomatology and, less frequently, by pharyngolaryngeal symptomatology.

Schwannomas may also arise on the cisternal portion of the nerves or on their retro-styloid pathway.

When large, their nervous origin may be difficult to determine, especially since the clinical symptomatology is not very characteristic.

It's therefore advisable to examine the effector organs clinically and by imaging: for example, hemiatrophy of the tongue (that should be looked for by MRI) indicates the diagnosis of schwannoma of CN XII.

In the imaging, as in the histology, the schwannomas of the lower cranial nerves have the same morphological appearance and signal as those of the vestibular schwannomas.

It consists of an oval or round mass, with distinct and regular contours, rather hypointense in T1, hyperintense in T2 with fairly intense enhancement. It may be heterogeneous with centro-tumoral zones of necrosis.

In the CT-scan, when located in the jugular foramen, it is accompanied by a regular enlargement of this foramen without aggressive lysis or major bone condensation reaction.

There is no neo-vascularization and arterial blush in the arteriography.

The treatment is surgery or radiation therapy (*gamma knife*).

The meningiomas [16]

Histologically, it most often consists of a meningothelial form (Fig. 9).

Clinically, the meningioma is not accompanied by a specific symptomatology. It may be related to impairment of CN IX and CN X (pharyngolaryngeal) or auditory or even minimally symptomatic.

In the imaging, the meningiomas are larger than thick, rather homogenous, with distinct and regular contours, rather homogenous. Their signal is hypointense in T1 and rather hyperintense in T2. This signal is highly enhanced after the injection of contrast product.

In the CT-scan, there is often an enlargement of the foramen with distinct condensing reaction.

In the arteriography, there is no arterial blush although there is considerable arterial vascularization.

The treatment is also surgical or radiation therapy.

The diagnostic range of these foraminal tumors comprises:

- for the jugular foramen: paraganglioma, schwannoma, meningioma;
- for the hypoglossal canal: schwannoma, meningioma.

Pathology of the retro-styloid space

Tumor pathology

The diagnostic range is the same as that of the jugular foramen with:

- schwannomas of CN IX, CN X, CN XI;
- vagal paragangliomas;
- meningiomas much more rarely.

A specific pathological frame: the carotid dissection

On the anatomo-pathological level, there is a contact in the retro-styloid space between the internal carotid artery inwards and the hypoglossal nerve outwards. Dissection of the internal carotid artery located in the superior cervical segment, will induce compression both of the cervical sympathetic ganglion and CN XII.

Local ischemic phenomena may also intervene to account for the impairment of CN XII in case of dissection of the internal cervical carotid artery.

In the imaging, the assessment of the association of paralysis of CN XII and a homolateral Horner's syndrome includes the search for carotid dissection with a T1-weighted sequence at best volume with fat saturation and an injected MR angiography to explore the supra-aortic trunks.

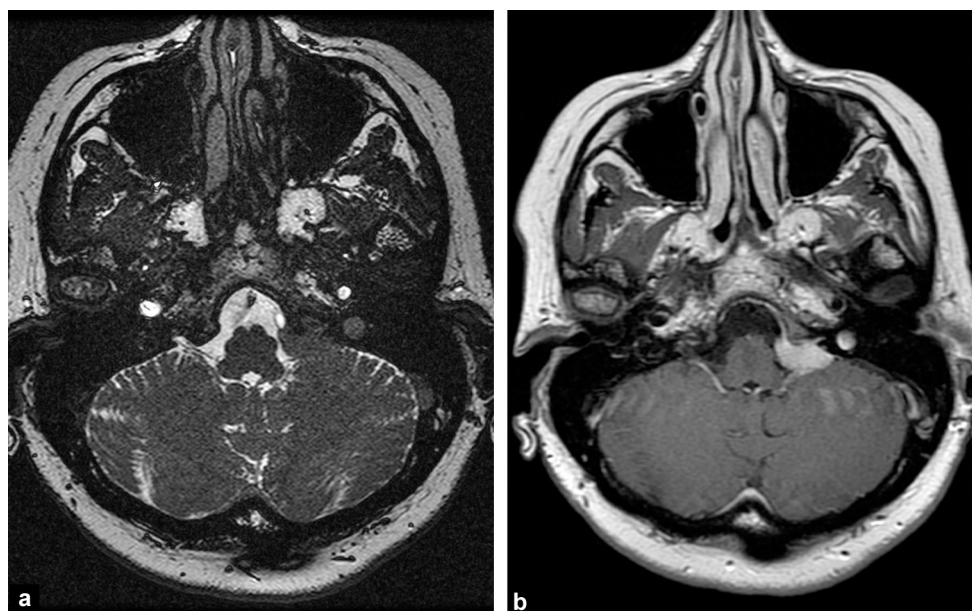


Figure 9. 49-year-old woman. Chance discovery: a: high-resolution T2-weighted axial slice; b: T1-weighted axial slice after injection of contrast product. Mass of the peribulbar cistern, extending to the left jugular foramen, enhanced after injection of contrast product, thicker than wide. Meningioma.

TAKE-HOME MESSAGES

- Only the glossopharyngeal (CN IX) and vagus (CN X) nerves are mixed (motor, sensory and secretory). The accessory (CN XI) and hypoglossal (CN XII) nerves are only motor.
- The diagnostic series of tumors of the jugular foramen is: paraganglioma, schwannoma, meningioma.
- The CT-scan is an important complementary tool for the MRI by studying the bone impairment of a tumor of the jugular foramen.
- The association of paralysis of CN XII and Horner's syndrome involves the search for a dissection of the internal cervical carotid artery by T1-weighted sequences with fat saturation and injected MR angiography.
- Glossopharyngeal neuralgia results from an artery-nerve conflict between CN IX and the posterior inferior cerebellar artery. It is manifested in the form of short and intense attacks of pain at the base of the tongue, the tonsils with irradiation towards the mandibular angle and the ear.
- Impairment of CN IX and CN X are, in most cases, concomitant.
- A lesion of CN XII will induce paralysis of all of the muscles of the tongue with visible atrophy in the imaging.

Clinical case

A 72-year-old woman. Increasing difficulty chewing (Fig. 10).

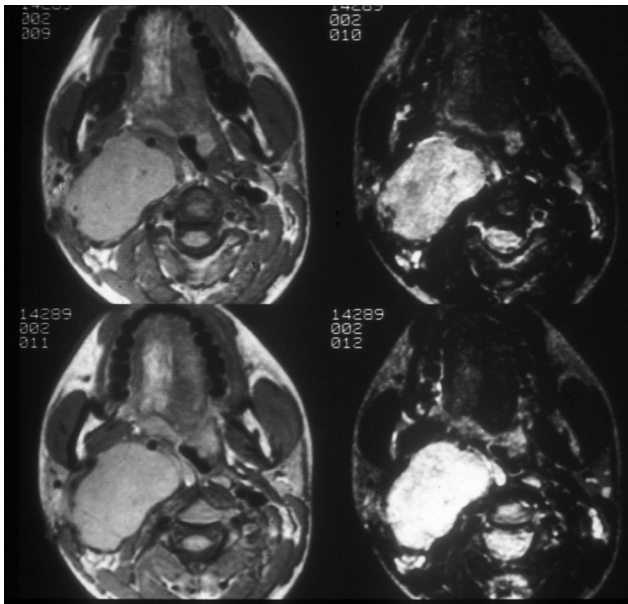


Figure 10. Left column: T1-weighted axial slices with injection. Right column: T2-weighted axial slices.

Questions

1. What is the most likely diagnosis for this mass?
2. What cranial nerve is involved in this disorder?

Answers

1. Oval mass of the retro-styloid space with distinct and regular contours, hyperintense in T2, enhanced after injection of contrast product. The most likely diagnosis is that of schwannoma.
2. There is atrophy of the right hemitongue, homolateral to the mass, attesting to impairment of the hypoglossal nerve (CN XII).

Diagnosis

Schwannoma of CN XII.

Message: always analyze the effector organs of the cranial nerves in particular, in case of suspicion of schwannoma.

Disclosure of interest

The authors have not supplied their declaration of conflict of interest.

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